

APPENDIX B
PENDING CLAIMS

52. (Three times amended) A method of detecting a lysosomal storage disorder (LSD), monitoring the progress of a LSD or the efficacy of treatment of a LSD in a human or animal subject, the method comprising assaying the level of a LSD marker in a biological sample derived from the subject, wherein the LSD marker is selected from the group consisting of Lamp-1 (lysosome-associated membrane protein type-1), 4-sulphatase and β -hexosaminidase, an increase in the level of the LSD marker in the subject relative to the corresponding level of the LSD marker in a non-affected individual or population being indicative of a LSD.

55. (Twice amended) The method according to claim 52, wherein the LSD marker is Lamp-1.

58. The method according to claim 52, wherein the biological sample comprises blood, plasma, urine, a fibroblast cell, a fibroblast cell culture or a fibroblast cellular extract.

59. (Once amended) The method according to claim 55, wherein the biological sample comprises blood, plasma, urine, a fibroblast cell, a fibroblast cell culture or a fibroblast cellular extract.

60. The method according to claim 59, wherein the biological sample comprises blood, plasma or urine.

61. The method according to claim 59, wherein the fibroblast cell or fibroblast cell culture is a skin fibroblast or skin fibroblast cell culture or a cellular extract thereof.

62. The method according to claim 61, wherein the fibroblast cell, fibroblast cell culture or fibroblast cellular extract is a Pompe, Salla, MPS II or MPS VI fibroblast cell, cell culture or cellular extract.

63. The method according to claim 52, wherein the LSD is selected from the list set forth in Table 1.

64. The method according to claim 63, wherein the LSD is selected from the group consisting of MPS I, MPS II, Gaucher disease, Pompe disease and Salla's disease.

65. (Once amended) The method according to claim 52, wherein the step of assaying the level of a LSD marker comprises measuring the enzyme activity of said LSD marker in the biological sample.

66. (Twice amended) The method according to claim 52, wherein the step of assaying the level of a LSD marker comprises contacting the biological sample with one or more antibodies specific for said LSD marker for a time and under conditions sufficient for the formulation of a complex to occur.

68. (Twice amended) The method according to claim 66, wherein the one or more antibodies are monoclonal antibodies.

69. (Once amended) The method according to claim 66, wherein the one or more antibodies is/are labeled with a reporter molecule.

70. (Once amended) The method according to claim 66, further comprising the step of contacting a complex formed between the LSD marker and one of the one or more antibodies with a labeled antibody for a time and under conditions sufficient for binding to occur.

71. (Once amended) The method according to claim 70, wherein the labeled antibody is labeled with a reporter molecule.

72. The method according to claim 69, wherein the reporter molecule is an enzyme, a fluorophore or a radionuclide molecule.

73. The method according to claim 72, wherein the enzyme, fluorophore or radionuclide molecule is selected from the group consisting of horseradish peroxidase, glucose oxidase, β -galactosidase, alkaline phosphatase, fluorescein, Eu^{3+} and other lanthanide metals, and rhodamine.

74. (Twice amended) The method according to claim 52, wherein:

- (a) the LSD is selected from the list set forth in Table 1;
- (b) the LSD marker is LAMP-1;
- (c) the biological sample comprises blood, serum or urine; and
- (d) the assay comprises measuring the enzymatic activity of the LSD marker or is an immunoassay.

93. (Twice amended) A method for detecting a lysosomal storage disorder (LSD), comprising assaying LAMP-1 in a sample of blood obtained from a patient that is asymptomatic for a LSD, an increase in the level of LAMP-1 in the patient relative to the corresponding level of LAMP-1 in a non-affected individual or population being indicative of a LSD.